

OPEN REDUCTION OF HIP DISLOCATION IN PATIENTS WITH ARTHROGRYPOSIS MULTIPLEX CONGENITA - AN ANTEROMEDIAL APPROACH

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ABSTRACT

Objective: To evaluate the results from surgical treatment of hip dislocation through the anteromedial approach, in patients with arthrogryposis multiplex congenita (AMC). **Methods:** The medical files and radiographs of seven children with AMC who presented hip dislocation (total of 10 dislocated hips) were retrospectively reviewed. Pre and postoperative joint mobility was evaluated by summing the joint range of motion in flexion and abduction. The acetabular angle and height of the femoral neck before the operation, and the continuity of the Shenton arc, Sharp angle and center-edge (CE) angle after the operation, were evaluated radiographically. When avascular necrosis was identified, it was classified in accordance with Ogden and Bucholz. **Results:** The mean age of the children at the time of the surgery was 5.5 months (range: 3 to 11 months). The

mean duration of follow-up for the patients was 9.5 years (range: 2 to 13 years). The mean amplitude of the sum of the joint range of motion in flexion and abduction in the preoperative examination was 108° (range: 70° to 155°) and postoperatively, it was 125° (range: 75° to 175°). In the last evaluation, eight hips were found to be centered and two were subluxated. Two hips had been subjected to Salter iliac osteotomy. Two hips (20%) had presented significant signs of Ogden type IV avascular necrosis. Eight hips had good results while two were fair. **Conclusion:** We consider that the anteromedial approach is a good option for treating hip dislocation in very young patients with arthrogryposis multiplex congenita.

Keywords – Arthrogryposis; Hip Dislocation; Surgical Procedures, Operative

INTRODUCTION

Arthrogryposis multiplex congenita (AMC) is a syndrome of unknown etiology and non-progressive nature that is characterized by the presence of multiple joint contractures with replacement of the musculature with fibrous bands and fat, limb deformity (generally symmetrical) and normal intelligence⁽¹⁾. Stern, in 1923, was the first to use this term (AMC), in descriptions

on four clinical cases from which he characterized the syndrome^(2,3).

The hip is affected in 51 to 85% of the cases of AMC, presenting fixed contractures and dislocation in 15 to 31% of the cases⁽⁴⁾.

Dislocation of arthrogrypotic hips is defined as a teratological condition, since it occurs at an early stage of intrauterine life. The acetabulum is small, shal-

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low and filled with fibrous-fatty tissue. The femoral head is hypoplastic and often flattened in its medial portion. The anteversion angle may vary greatly and may be retroverted, unlike the typical hip dislocation, in which the anteversion angle is greater in the great majority of cases^(5,6).

Arthrogryptic hips are characterized by diminished mobility, absence of normal muscle function and predisposition towards loss of mobility after surgical treatment^(5,7,8). These characteristics create difficulty in dealing with these hips. Closed reduction is generally ineffective^(3,9). Some authors have recommended that bilateral hip dislocation should remain untreated because of the high incidence of complications^(6,7,9,10), although this is a matter of controversy⁽¹¹⁾. In cases of unilateral dislocation, most authors have indicated surgical treatment, with the aim of avoiding pelvic tilt and scoliosis^(6,7,11-13).

Reduction of the dislocation by means of an anterior iliofemoral approach associated with femoral shortening is the procedure most used among children over the age of 18 months, although some authors have performed this on very young children through an anteromedial approach, with satisfactory results^(4,14).

The present study had the aim of evaluating the results from surgical treatment to reduce hip dislocation through an anteromedial approach, among patients with AMC.

MATERIALS AND METHODS

The medical files and radiographs of 51 patients who presented AMC were reviewed. Fourteen patients presented 23 dislocated hips. Between January 1990 and January 2006, 10 dislocated hips in seven children were reduced surgically by means of the anteromedial approach, at Hospital Pequeno Príncipe.

AMC was diagnosed in accordance with Fisher's clinical criteria: 1) joint contractures at birth, in at least two joints, except for congenital club foot; 2) absence of progressive neurological disease; 3) evidence of muscle weakness with fusiform joints; presence of 'dimples' (skin folds) or pterygium, or absence of skin folds⁽¹⁾.

The subjects of this study were cases with a minimum follow-up of 24 months that had not undergone previous treatment.

The degree of functioning was not used as a criterion for classifying the results. However, all the children were classified postoperatively as community walkers, home

walkers, non-functional walkers or non-walkers⁽¹⁵⁾. The patients' requirements for the use of locomotion aids were recorded.

The results were evaluated in relation to the clinical and radiographic data.

The pre and postoperative clinical evaluation consisted of the sum of the hip range of motion in flexion and abduction. The results were considered to be good, fair or poor according to the degree of joint mobility at the last consultation and the presence or absence of hip subluxation (Table 1).

Table 1 – Table of criteria for evaluating the results.

	Range of motion in flexion + abduction	Radiograph
Good	≥ 90°	Centered
Fair	60°- 89°	Subluxated
Poor	Stiffness	Dislocated

In the preoperative radiographic evaluation, the acetabular angle and height of the proximal femur in relation to the Hilgenreiner line were measured.

In the postoperative evaluation, measurements of the acetabular angle were used for children whose triradiate cartilage was still open, while measurements of the Sharp angle⁽¹⁶⁾ were used for children who triradiate cartilage was closed. The Shenton arc integrity and Wiberg center-edge (CE) angle⁽¹⁷⁾ were also evaluated in children over the age of five years. Hips in which the Shenton arc was complete and the CE angle was greater than or equal to 15° were considered to be centered. Avascular necrosis was classified in accordance with Ogden^(18,19).

RESULTS

Among the seven patients who participated in this study, four were male and three were female. Three patients presented bilateral hip dislocation and four had unilateral dislocation (two on the right side and two on the left side).

The children's mean age at the time of the surgery was 5.5 months (range: three to eleven months). The patients were immobilized for a six-week period and an Ilfeld brace was used on three patients after the plaster cast had been removed (Table 2). The mean duration of the follow-up was 9.5 years (range: two to thirteen years).

The mean hip range of motion in flexion and exten-

sion in the preoperative examination was 92.5° (range: 60° to 135°) and the mean range of motion in abduction was 19° (range: 0° to 30°). After summing the preoperative flexion and abduction ranges, the mean was found to be 108° (range: 70° to 155°).

Table 2 – Evaluation of the patients according to sequential number and initials, side affected, age at the time of treatment, duration of follow-up, preoperative acetabular angle, last acetabular angle or Sharp angle, last Shenton arc broken (B) or intact (I), preoperative level of proximal metaphysis of the femur, and CE index at the last consultation.

Patient	Side affected	Age at time of treatment	Duration of follow-up		Lat acetabular angle or Sharp(S) angle	Last Shenton arc	Preoperative level of proximal metaphysis	CE
1.	R	6 m	13 y + 8 m	50°	S = 49°	B	0	7°
2.	L	6 m	13 y + 8 m	45°	S = 45°	I	0	17°
3.	L	7 m	13 y + 2 m	42°	S = 48°	I	0	40°
4.	L	11 m	5 y + 4 m	36°	S = 38°	I	0	0°
5.	R	3 m	9 y + 1 m	40°	S = 42°	I	0	46°
6.	L	3 m	9 y + 1 m	45°	S = 42°	I	0	46°
7.	R	8 m	14 y + 3 m	28°	S = 48°	I	0	32°
8.	R	4 m	2 y + 10 m	33°	22°	I	-1	-
9.	L	4 m	2A + 10	45°	20°	B	0	-
10.	R	3 m	5A+4	45°	21°	I	-1	

The mean postoperative hip range of motion in flexion and extension 92° (range: 50° to 130°), while the mean range of motion in abduction was 33.5° (range: 10° to 65°). After summing the postoperative flexion and abduction ranges, the mean was found to be 125° (range: 75° to 175°). Two hips presented a summed range of motion of between 60° and 89° (fair), eight hips showed ranges greater than 90° (good) and there were no cases of joint stiffness (poor) (Table 3).

Regarding radiographic evaluations, the acetabular angle was measured in the preoperative examination, and the mean value was found to be 41° (range: 28° to 50°). Eight out of the ten hips presented the proximal metaphysis of the femur at the level of the Hilgenreiner line.

After the operation, the mean acetabular angle was 21° (range: 20° to 22°) in three hips. The mean value of the Sharp angle in seven hips was 44.5° (range: 38° to 49°). The mean CE angle was 26.8° (range: 7° to 46°) in seven hips of children over the age of five years (Table 2).

In relation to function, six patients were community walkers and one was a non-walker. Three patients were able to walk without aids and three were able to walk with bilateral braces: five of ankle-foot type and one of knee-ankle-foot type. The patient who was a non-walker

made use of knee-ankle-foot braces bilaterally.

Out of the ten hips that were reduced by means of the anteromedial approach, eight of them were found to have become centered at the last consultation and two presented subluxation. Type IV avascular necrosis was

observed in two hips, while the other eight hips did not present necrosis (Table 4).

One patient presented subluxation in both hips (Figure 1). Right-side Salter iliac osteotomy was performed two years and six months after the hip reduction, and the same procedure was performed on the left side two years and eleven months after the anteromedial hip reduction (Figure 2). At the last evaluation, the left hip was still subluxated (Figure 3 and Table 4; case two).

DISCUSSION

Only limited information is available in the literature regarding the treatment for hip dislocation in cases of AMC in patients under the age of two years, and the existing studies present small numbers of patients^(8-10,20,21). Most of the studies described in the literature focused on total patient management, without much specific or consensual data on treatments for hip dislocation⁽²²⁾.

Regarding the reduction methods for hip dislocation, the literature is consistent in that most authors believe that closed reduction is ineffective^(5,8,13,20,23). Within a series described by Gruel et al⁽¹³⁾, they treated two patients by means of closed reduction and obtained poor results, while better results were obtained through open reduction using the anterior iliofemoral route with femo-

Table 3 – Distribution according to initials, preoperative abduction, last abduction, preoperative flexion, last flexion, summed preoperative range of motion and summed postoperative range of motion.

Patient	Preoperative abduction	Last abduction	Preoperative flexion	Last flexion	Preoperative sum	Postoperative sum
	Pré	ult	Pré	Ult	Pré-Op	Pós-Op
1.	10°	25°	60°	50°	70	75
2.	15°	30°	60°	60°	75	90
3.	20°	20°	90°	70°	110	90
4.	30°	40°	120°	110°	150	150
5.	30°	40°	-	130°	-	170
6.	30°	40°	-	130°	-	170
7.	20°	40°	135°	90°	155	130
8.	-	10°	-	70°	-	80
9.	-	25°	-	100°	-	125
10.	0°	65°	90°	110°	90	175
Results	19,3°	33,5°	92,5°	92°	108,3	125,5

Table 4 – Distribution according to initials, general registration, date of birth, sex, final result and Ogden necrosis classification.

Patient	Registration	Birth (dd/mm/yyyy)	Sex	Results	Ogden
1.	5238	17/07/1991	M	Centered	4
2.	5238	17/07/1991	M	Subluxated	-
3.	35305	21/01/1992	M	Centered	4
4.	57124	11/03/2000	F	Centered	-
5.	47444	17/06/1996	M	Centered	-
6.	47444	17/06/1996	M	Centered	-
7.	30116	15/09/1990	M	Centered	-
8.	2241391	30/05/2003	F	Centered	-
9.	2241391	30/05/2003	F	Subluxated	-
10.	57140	28/09/1999	F	Centered	-



Figure 1 – Cases 1 and 2: preoperative arthrography.



Figure 2 – Case 1, twenty months after the operation: subluxation of the right hip.

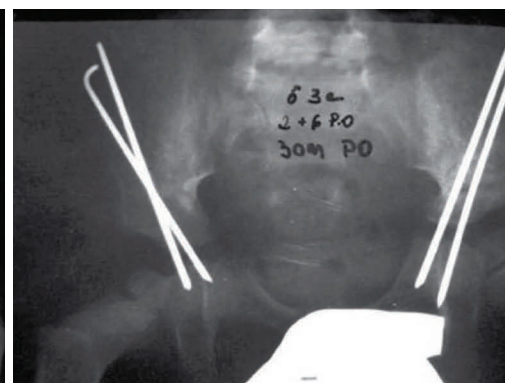


Figure 3 – Cases 1 and 2, thirty months after the operation: bilateral Salter iliac osteotomy.

ral shortening. In the present study, the closed reduction technique was not used on any of the dislocated hips.

In the same way as in cases of congenital hip dislocation, age influences the choice of approach. Sta-

heli *et al*⁽¹¹⁾ and Herring and Banta⁽¹⁴⁾ advocated that open reduction via the anteromedial route should be used up to the age of six months, although Coleman⁽⁵⁾ mentioned that this method could be used up to the

age of two years.

For arthrogryptic hips, the surgical approach that is recommended in cases over the age of two years is surgical reduction by means of the anterior iliofemoral approach, with femoral shortening, with the aim of facilitating the reduction and diminishing the incidence of avascular necrosis⁽³⁾.

The reports in the literature show that there is a relatively frequent need for secondary procedures associated with open reduction of arthrogryptic hip dislocation, especially when the anteromedial access route is used, because of progressive subluxation^(4,8,10,13). In the present study, secondary procedures were needed in the cases of two hips that were reduced by means of the anteromedial approach. Staheli et al⁽¹¹⁾ reported that the results relating to range of motion in the hips treated with anteromedial reduction were better than in the hips that were reduced using the iliofemoral route because capsulorrhaphy was not performed.

In our study, we could see that all of the ten hips treated with reduction via the anteromedial route gained range of motion, with eight good results and two with fair results because of continued subluxation despite the lack of clinical symptoms.

There is a consensus in the literature that unilateral hip dislocation in children up to the age of two years who present arthrogryposis should be treated to prevent the development of pelvic tilting, leg length discrepancy and, secondarily, scoliosis. The preferred procedure is reduction by means of the iliofemoral approach, with proximal femoral shortening^(6,7,11-13).

Some authors have reported good results through anteromedial reduction, and they recommended this route up to the age of six months, even when both hips were affected^(5,10,11,14).

Szöke et al⁽⁴⁾ recommended open reduction using the anteromedial route in unilateral and bilateral cases between the ages of three and six months, in combination with other surgical corrections for congenital contractures of the knees and feet.

The management for the hips in patients with AMC who present bilateral dislocation is very

controversial^(11,13), and the indications for treatment are not very clear.

Gibson and Urs⁽⁹⁾, Williams⁽¹⁰⁾ and Shapiro and Bresnan⁽²¹⁾ reported that bilateral dislocation should not be treated, because the hips do not present instability or pain over the course of the evolution, whereas after the treatment, joint stiffness and pain could develop. Gruel et al⁽¹³⁾ did not treat bilateral hips when the patient did not present a prognosis of becoming able to walk, and we agree with this.

St. Clair and Zimble⁽²²⁾ indicated treatment for cases of bilateral dislocation in which the upper limbs were mildly affected and the hips had a good range of motion. However, some authors have reported performing surgical reduction treatment for bilaterally dislocated hips, independent of whether the upper limbs were affected^(4,5,11,13,14,22).

Staheli et al⁽¹¹⁾ reported on 18 patients (24 hips) with AMC and hip dislocation. The hips treated with open reduction using the anteromedial route presented better mobility than did those treated with closed reduction or with open reduction using the anterior iliofemoral route, with good results: good in 80% of the cases; fair in 12%; and poor in 8%^(2,11). Based on our study, good results are obtained when dislocated hips are treated by means of the anteromedial approach, among children with AMC under the age of one year who present unilateral or bilateral dislocation.

In cases of older children who present unilateral dislocation, surgical reduction associated with femoral shortening is the preferred treatment⁽³⁾.

CONCLUSION

Although this study had a small sample of cases, we observed that none of them evolved with joint stiffness, that the anteromedial technique did not diminish the joint mobility, and that it was possible keep the hip centered in most cases. Thus, we consider that the anteromedial route is a good option for treating unilateral or bilateral hip dislocation in patients with AMC within their first year of life who present potential for gaining the ability to walk, independent of upper-limb function.

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